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Neonatal Ebstein's Anomaly

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Abstract

Ebstein's anomaly is a congenital heart disease that results from failure of delamination of the tricuspid valve with resulting apical displacement of the septal and posterior leaflets of the tricuspid valve. Age at presentation can vary greatly but neonatal presentation is associated with extraordinary high mortality rates. Comprehensive multispecialty care is required starting at the time of fetal diagnosis. Fetal echocardiography is vital in monitoring progression of the disease in utero. Fetal echocardiogram can evaluate for complications such as arrhythmias, pericardial effusion, or fetal hydrops. Post-natal evaluation should include evaluation of functional pulmonary atresia or circular shunt. Despite advances in surgical technique for Ebstein's anomaly, mortality for it remains high with early surgical intervention. Aggressive medical management should be used to support patients with Ebstein's anomaly during the neonatal period. Surgical procedures for neonatal Ebstein's vary widely from systemic to pulmonary shunts with or without tricuspid valve closure to tricuspid valve repair.

Keywords: neonatal Ebstein's anomaly, Ebstein's anomaly, tricuspid valve dysplasia, Fetal Ebstein

1. Introduction

Ebstein's anomaly (EA) was first described by Wilhelm Ebstein in 1866 noting the septal and inferior leaflets of the tricuspid valve arose from the right ventricular myocardium [1]. EA is a rare congenital heart disease with a prevalence of 2.4 per 10,000 live births [2]. Embryologically, EA is a result of varying degrees of failure of leaflets to delaminate from the underlying endocardium that results in a number of characteristic features. There are varying degrees of apical displacement of the tricuspid leaflets with the septal leaflet most severely affected followed by the posterior leaflet. Furthermore, the right ventricle (RV) is myopathic and is separated into

two zones, with an “atrialized” poorly functional portion between the true annulus and the hinge point of the apically displaced septal leaflet, while the “functional” RV is the portion below the leaflet hinge point. Depending on the degree of leaflet displacement this functional RV volume can be quite diminutive.

The clinical manifestations of EA vary widely from mild forms presenting in adulthood, to severe forms with high mortality in the neonatal period. In utero, EA can result in hydrops and arrhythmia. Furthermore, an in utero diagnosis of EA has an incidence of 48% fetal demise [3]. Mortality rates are highest in the neonatal period ranging from 17 to 56% [4] and poses significant medical and surgical challenges.

2. Associated anomalies and arrhythmias

Ebstein anomaly is known to have several additional cardiac manifestations. A patent foramen ovale or atrial septal defect is normally present. The right to left shunting across the defect accounts for the relative hypoxia exhibited in certain patients. RV outflow tract obstruction in the form of anatomical pulmonary atresia occurs in about half of the symptomatic neonates requiring surgical intervention [5]. In the setting of pulmonary atresia, a patent ductus arteriosus is required as the source of pulmonary blood flow [6–8]. Left ventricular non-compaction cardiomyopathy has been noted to be associated with EA. A retrospective study demonstrated that 10 of 61 patients (16%) with EA also had left ventricular non-compaction [7]. This was associated with a higher mortality risk of 30% in those with LVNC compared to 13% with EA alone. Wolff-Parkinson-White (WPW) is present in about 10–30% of cases. In about 20% of cases with WPW, there may be more than two accessory pathways present. The accessory pathways are usually present on the tricuspid valve annulus [9, 10]. Due to large right atrium, EA patients are at risk for atrial tachycardia, atrial flutter, intra-atrial reentrant tachycardia, atrial fibrillation, AV node reentrant tachycardia, and ventricular arrhythmias.

3. Pathologic anatomy

Carpentier et al. described the characteristic features of this disorder that are relevant to surgical management [11].

1. Failure of delamination of the TV leaflets is the hallmark of EA whereby the leaflets are tethered to the endocardium by fibromuscular attachments or abnormal foreshortened chordae. Each leaflet exhibits varying degrees of apical displacement and tethering with the septal leaflet most severely affected followed by posterior then anterior leaflets. This results in anterior and apical displacement of the functional annulus.
2. The anterior leaflet is attached to the true anatomical annulus but is large or sail like.

3. The portion of the RV above the functional annulus ('atrialized right ventricle') is dilated and thin. The true tricuspid annulus is almost always enlarged. In a neonate this measures approximately 21 mm.
4. The cavity of the effective RV is reduced ('functional right ventricle').
5. The infundibulum of the RV can be obstructed by the redundant tissue of the anterior leaflet and its chordal attachments to the infundibulum.

In addition to the leaflet abnormalities there is a variable degree of ventricular myocardial dysfunction. Morphometric histopathologic studies have demonstrated that there is an absolute decrease in the number of myocardial fibers in addition to thinning of the wall of the dilated RV in EA [12].

Carpentier et al. also described four grades of Ebstein's anomaly [11].

Type A: The anterior leaflet has normal morphology and the RV is adequate.

Type B: The anterior leaflet has abnormal chordae but normal mobility. The RV is reduced in volume but adequate.

Type C: The anterior leaflet is restricted in movement. The RV is small with a large atrialized component.

Type D: Also called 'tricuspid sac' as the leaflets form a complete sac of fibrous tissue adherent to the RV. The only functional part of the RV is the infundibulum.

3.1. Perinatal period

The long term prognosis of a fetus diagnosed with EA is poor and remains one of the highest mortalities amongst congenital heart disease patients. One multicenter study showed that a fetal diagnosis of EA resulted in a 17% fetal demise. Furthermore, there was an additional 32% in-hospital attrition of live-born babies with EA with an overall 45% perinatal mortality [4]. Risk factors for perinatal mortality include lack of antegrade flow across the pulmonary valve, large cardiothoracic ratio, earlier in utero diagnosis, large tricuspid valve annulus, pericardial effusion, and right ventricular dysfunction [13–16]. Pulmonary valve regurgitation may be the most ominous risk factor representing the end result of severe tricuspid regurgitation with resultant volume load on a myopathic right ventricle that has to pump against retrograde flow from the PDA. This triad of diminished preload, increased afterload and a dysfunctional right ventricle leads to inadequate preload to the left ventricle and subsequent heart failure, cardiogenic shock and perinatal demise. These factors in-utero lead to hydrops and arrhythmias and ultimately fetal demise.

4. Pathophysiology of the newborn

Neonates are symptomatic as a result of ineffective RV cardiac output and severe TV regurgitation. There is usually some degree of cardiomegaly which can be quite severe compressing

the lungs. Furthermore, cyanosis results from systemic venous return being shunted across the ASD to the left side of the heart. Neonatal pulmonary vascular resistance (PVR) is elevated and this is a major impediment to effective antegrade flow from the diminutive and myopathic RV. In the first week of life when pulmonary vascular resistance is high pulmonary blood flow is dependent upon the PDA. This results in a physiological state referred to as “functional” pulmonary atresia. When the PVR decreases over the first week of life, the RV may then be able to overcome the afterload to establish antegrade flow. True anatomical pulmonary atresia where there is luminal discontinuity between RV and pulmonary artery is also often seen in these symptomatic neonates. These patients will have ductal dependent pulmonary circulation until a surgical procedure is performed to establish pulmonary blood flow. Left ventricular dysfunction can also play a critical role in the development of decompensated heart failure. This is related to left ventricular displacement of the interventricular septum as a result the severely dilated dysfunctional RV. This “pancaking” of the LV cavity impedes filling and diminishes systemic cardiac output. In less severe forms of EA the RV can generate effective antegrade flow especially when the PVR decreases. Antegrade flow across the RV outflow tract is accompanied by clinical improvement in symptoms. Neonates with severe TR or gross cardiomegaly who are otherwise asymptomatic have an associated mortality of 45% within the first year of life without intervention [17, 18]. The natural history of being diagnosed with EA during infancy is sobering [19]. However those who survive early childhood can expect reasonable longevity. When the disease is mild symptoms are not noticed until later in adult life. Symptoms are often related to exercise intolerance or cyanosis from progressive tricuspid regurgitation.

5. Diagnostic evaluation

5.1. Chest X-ray

Depending on the severity of disease, the chest roentgenograms usually demonstrates massive cardiomegaly with decreased pulmonary vascular markings (**Image 1**).

5.2. Electrocardiography

The electrocardiograms for patients with EA are usually abnormal. The most common finding on ECG are tall P waves and right bundle branch block. The tall P waves are indicative of a large right atrium. The right bundle branch block occurs because abnormal development of the right bundle branch which appears to be associated with septal leaflet and medial papillary muscle development on necropsy studies [5]. Some patients may have a prolonged PR interval from long intra-atrial conduction times from a large right atrium. Wolff-Parkinson-White syndrome is associated with EA, thus ventricular pre-excitation may be seen on ECG.

5.3. Echocardiography

Echocardiography is the gold standard for obtaining the diagnosis for EA. Two dimensional (2-D) echocardiography can evaluate the tricuspid valve leaflets and their excursion. The apical four

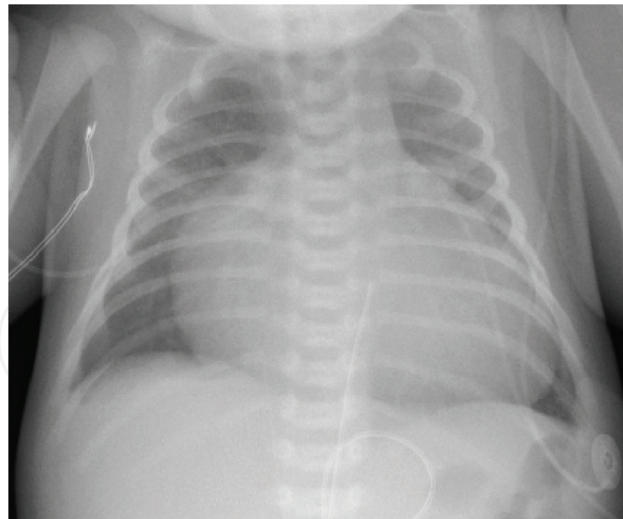


Image 1. Chest x-ray of an infant with EA. there is marked cardiomegaly with a significant cardiothoracic ratio. With marked cardiomegaly lung development can be impaired.

chamber views allow calculation of the displacement index, which measures the distance from the true septal annulus to the level of the apically displaced septal leaflet hinge point (**Image 2**). Distance is indexed to body surface area and values $>8 \text{ mm/m}^2$ are consistent with EA [20]. Color Doppler echocardiography can demonstrate the presence and location of tricuspid valve regurgitation (**Image 3**) [21]. However, severity can be difficult to quantitate due to apical displacement. RV dysfunction and functional or anatomic pulmonary atresia can be evaluated by 2-D and color echocardiography [21] **Image 4**. The Great Ormond Street Echocardiogram (G.O.S.E.) score is a mortality risk stratification score for neonates with EA. It is calculated from the apical four chamber view by adding the right atrium and atrialized right ventricular volume and dividing by the sum of the functional right ventricular volume, left atrial and ventricular volumes. (18) A G.O.S.E score of 3 (ratio of 1.1–1.4) with cyanosis or 4 (ratio > 1.5) has a mortality of nearly 100% [18] **Image 5**.

Echocardiography can define other associated abnormalities with EA such as the presence of a patent ductus arteriosus, size and direction of shunting through the atrial septal defect/patent foramen ovale, presence of a ventricular septal defect, and hypertrabeculated left ventricle suggesting left ventricular non-compaction cardiomyopathy.

Fetal echocardiography is a useful diagnostic tool for prenatal diagnosis and monitoring progression of disease in utero. The 4-chamber view of the fetal heart will demonstrate apical displacement of the tricuspid valve annulus, enlarged right ventricular and atrial size, and large tricuspid valve annulus (**Image 6**). Color flow imaging can be used to evaluate the degree of tricuspid valve regurgitation. The pulmonary valve can be evaluated by 2-D and color flow imaging to assess for pulmonary atresia. M-mode assessment can determine any rhythm abnormalities such as supraventricular tachycardia [22]. In addition, signs of hydrops such as pericardial effusions can be visualized (**Image 7**).

Fetal echocardiogram is important for monitoring clinical status of the fetus during pregnancy. A large multicenter study performed by Freud et al. evaluated over 400 fetal echocardiograms of patients with EA. They demonstrated that larger cardiothoracic ratio, more than moderate

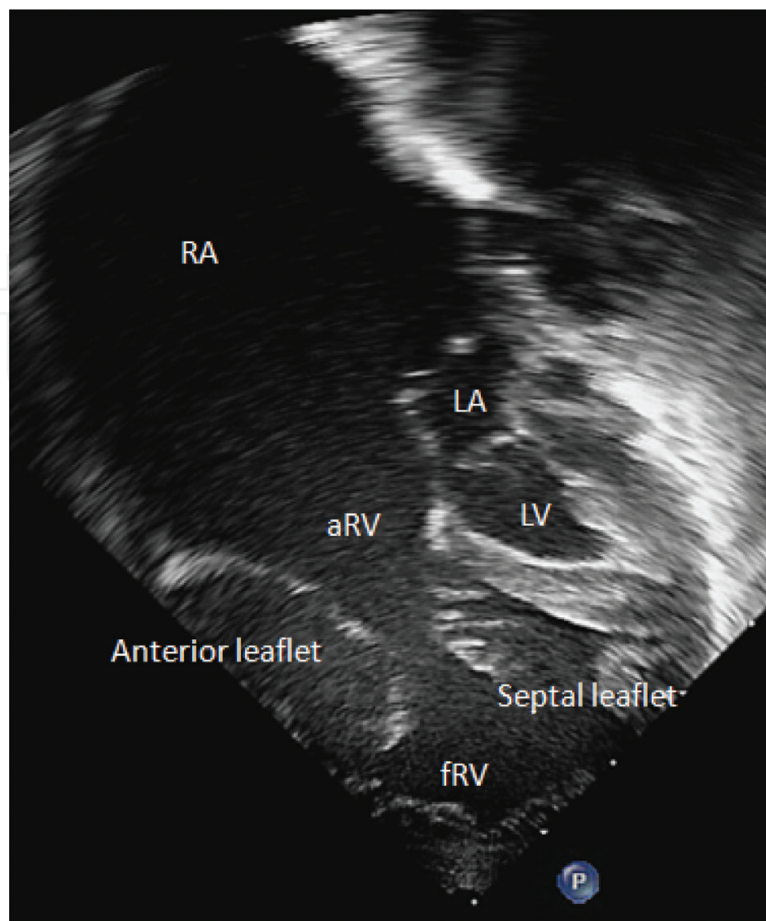


Image 2. Apical 4 chamber view of Ebstein's anomaly with severe enlargement of the right atrium and atrialized portion of the right ventricle. There is apical displacement of the tricuspid valve with tethering of the septal leaflet leading to poor coaptation and tricuspid valve regurgitation. RA: Right atrium. LA: Left atrium. aRV: Atrialized right ventricle. LV: Left ventricle. fRV: Function right ventricle.

tricuspid regurgitation, larger tricuspid annulus diameter z-score, larger diameter vena contracta for tricuspid regurgitation, lack of antegrade pulmonary blood flow, pulmonary regurgitation, and pericardial effusion were associated with increased perinatal mortality [4]. Furthermore, Tierney et al. demonstrated that only 31% of fetuses had no predictive risk factors for poor hemodynamic status at time of diagnosis, and of those, 61% went on to develop one or more signs later in gestation. As such, frequent fetal echocardiograms are necessary to monitor the clinical status of fetuses with EA [23].

5.4. Computed tomography/magnetic resonance imaging

There is limited utility for the use of CT, MRI, or cardiac catheterization in neonatal EA.

5.5. Treatment

5.5.1. Medical

In a study of 415 neonates presenting with symptomatic EA the overall hospital mortality was 24% [19]. Furthermore, surgical intervention in the neonatal period across US hospitals is

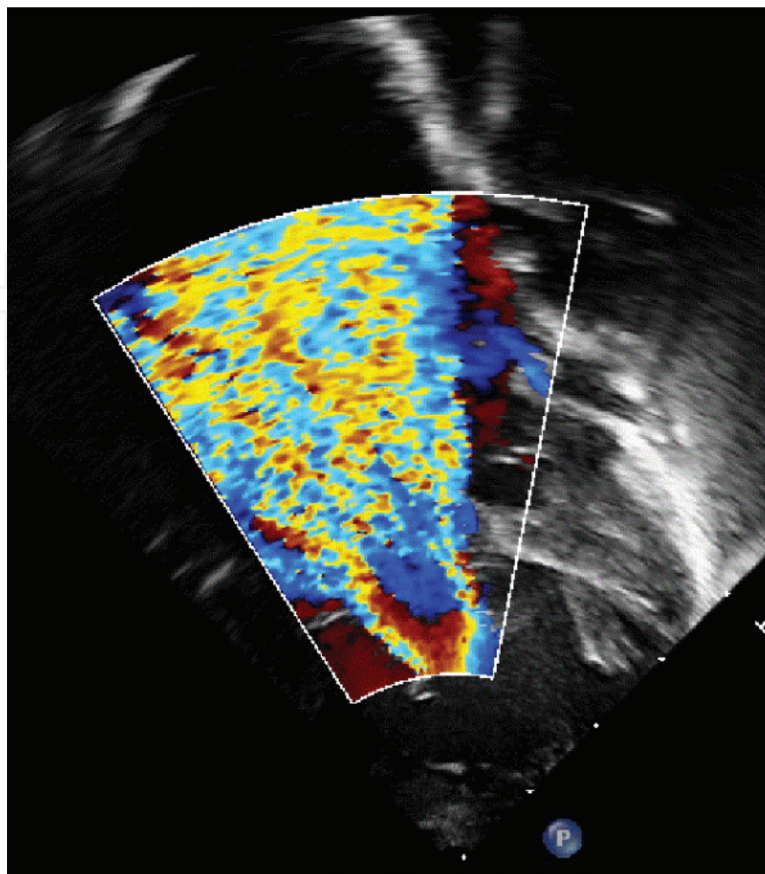


Image 3. Apical 4 chamber with color flow imaging of the tricuspid valve. There is severe tricuspid valve regurgitation due to poor coaptation of the tricuspid valve leaflets.

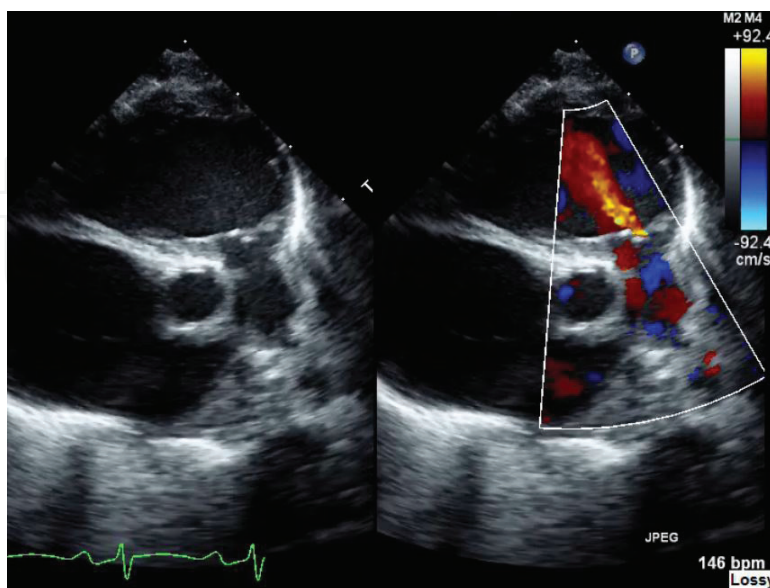


Image 4. Color compare parasternal short axis view demonstrates the small functional right ventricle and pulmonary atresia. The tricuspid valve is rotated in a fashion that the effective orifice opens to the right ventricular outflow tract. Color Doppler demonstrates swirling of blood in the functional right ventricle, tricuspid regurgitation, and no antegrade pulmonary blood flow. aRV: Atrialized right ventricle.

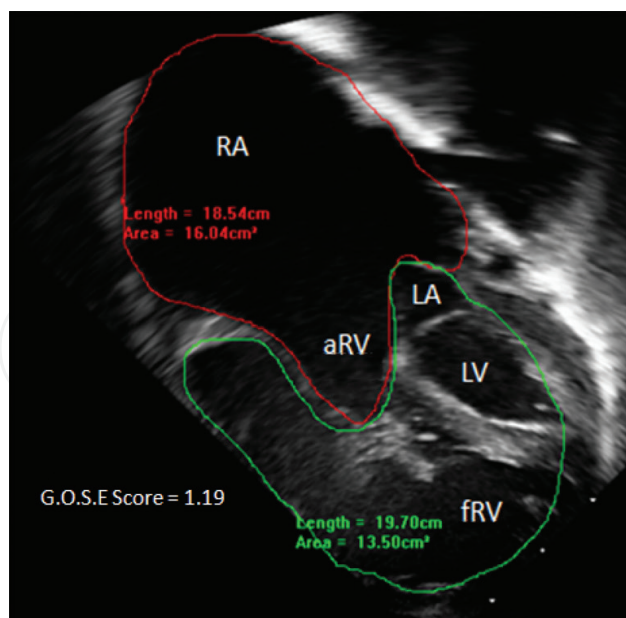


Image 5. Apical four chamber view demonstrating the right atrium, atrialized portion of the right ventricle, functional right ventricle, left atrium and left ventricle. There is a grade 3 GOSE score which correlates with 100% mortality in presence of cyanosis. RA = right atrium. aRV = atrialized right ventricle. fRV = functional right ventricle. LA = left atrium. LV = left ventricle.

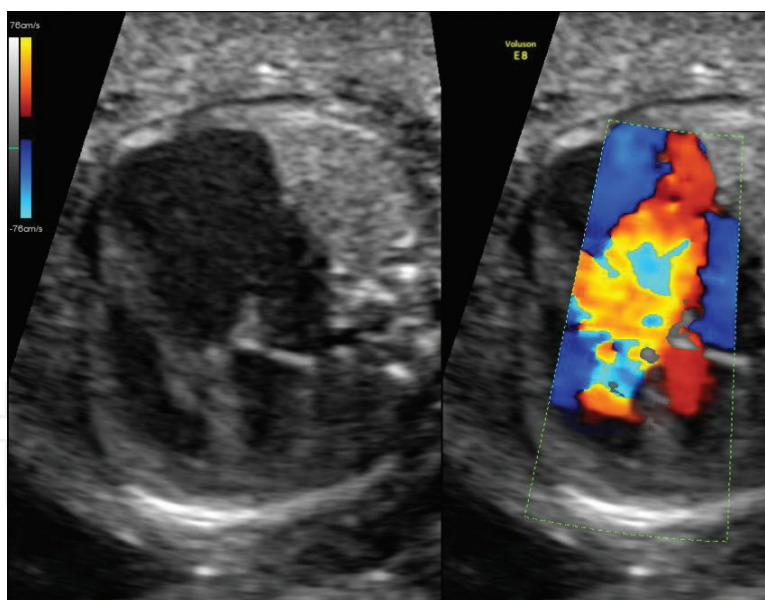


Image 6. Fetal echocardiogram performed at 22 weeks and 3 days gestation. There is severe enlargement of the right atrium with severe tricuspid valve regurgitation demonstrated on color Doppler evaluation. The tricuspid valve annulus dimension is markedly enlarged.

associated with a mortality of 27–36% depending on the procedure performed [19]. There is significant improvement in surgical outcome if the patient can be medically managed out of the neonatal period [24]. As such the best survival rates for EA occurs outside of the neonatal period, thus medical management with supportive care is crucial for improving outcomes.

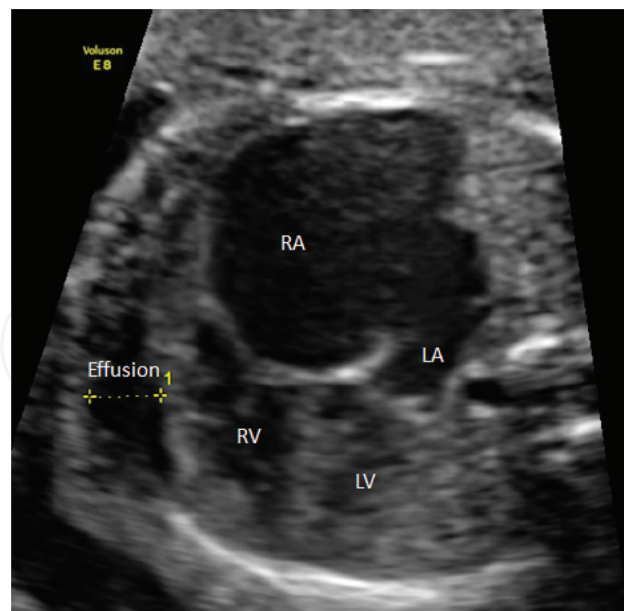


Image 7. Fetal echocardiogram at 22 weeks and 3 days gestation with a four chamber view of the heart. The heart mass takes up the entire thoracic cavity. There is severe enlargement of the right atrium with evidence of a pericardial effusion. RA: Right atrium. RV: Right ventricle. LA: Left atrium. LV: Left ventricle.

Relatively stable but symptomatic patients can be treated with prostaglandin infusion to maintain ductal patency if functional or anatomical pulmonary atresia is evident. Supplemental oxygen should not be greater than 21% fractional inspired oxygen to avoid pulmonary overcirculation and volume loading of the heart. During this phase oxygen saturation should be maintained between 75 and 85%. As the pulmonary vascular resistance decreases, prostaglandin therapy can be discontinued. This will also allow for proper evaluation of antegrade pulmonary blood flow as the ductus closes. If saturations decrease under 80% then agents to lower pulmonary vascular resistance can be administered to promote antegrade flow, these include supplemental oxygen and nitric oxide.

Use of prostaglandins in the presence of pulmonary valve regurgitation may exacerbate heart failure symptoms due to the development of a circular shunt. In this physiology blood flows from left ventricle to aorta, then it is shunted away from the systemic circulation via the large ductus to the pulmonary artery then retrograde via the pulmonary valve into the right ventricle to right atrium via the incompetent tricuspid valve then back to the left side of the heart via the ASD. This creates high output heart failure. Prostaglandins should be stopped if this physiology exists.

Further hemodynamic instability leads to cardiogenic shock. In these situations intubation and mechanical ventilation with large tidal volumes are key to promoting adequate ventilation in patients with massive cardiomegaly. Furthermore, sedation and possible paralysis to limit oxygen requirements may be required. Inotropic support in the form of milrinone complemented with low dose of dopamine or epinephrine may be necessary to assist with cardiac output. Tachyarrhythmias are common in this group of patients so utilization of catecholamine inotropes should be used sparingly.

Milrinone is a very effective drug since it has lusitropic and inotropic effects on the right ventricle. Furthermore, it decreases the pulmonary vascular resistance which promotes antegrade pulmonary blood flow. Frequent echocardiograms during the first week are useful to assess antegrade flow across the RV outflow tract and degree of TV regurgitation. This assessment will help guide weaning prostaglandins, and initiation of nitric oxide and inotropes.

In summary, medical management when pulmonary ductal dependency exists is analogous to single ventricle physiology whereby a balance between systemic and pulmonary circulation needs to be established. This is best done with maintenance of prostaglandins and low oxygen supplementation. Once the pulmonary vascular resistance drops and there is antegrade flow across the pulmonary valve then this management is more analogous to two ventricle physiology with a poor right ventricular pump. As such this is best managed by stopping prostaglandins and allowing for ductal closure. Concomitantly administration of nitric oxide, milrinone and higher oxygen supplementation will augment antegrade flow.

The goal of medical therapy is to avoid an operation particularly during the neonatal period when mortality is highest for any surgical procedure performed.

5.6. Surgical indications

Surgical indications for EA include failure to wean from mechanical ventilator support, failure to wean off prostaglandin with systemic oxygen saturation below 75%, functional or anatomic pulmonary atresia, Great Ormond Street Echocardiography (G.O.S.E.) score of 3 or 4, and patients with right heart failure.

5.7. Surgical procedures

To date there has been many procedures described for the surgical treatment for neonatal EA [11, 25–28, 31–35].

Danielson at the Mayo Clinic first described some of the essential principles for EA repair in any age group [25]. This includes plication of the atrialized RV, posterior tricuspid annuloplasty, closure of ASD and right reduction atrioplasty.

The cone reconstruction first described by da Silva and colleagues has now evolved into the technique of choice when repairing the tricuspid valve for EA [27]. In this procedure, the anterior and posterior leaflets of the tricuspid valve are mobilized from their attachments to the RV endocardium maintaining free edge attachments. The mobilized leaflets are rotated clockwise and then reattached to the true annulus [36]. To date there has been a growing number of reports of utilization of the Cone procedure for neonatal EA [27].

Starnes et al. reported a single ventricle palliation strategy for neonates with good outcome [29]. In the Starnes procedure the RV is excluded by performing a fenestrated patch closure of the tricuspid valve. An atrial septectomy allows for excellent mixing. Finally, a modified Blalock-Taussig-Thomas shunt is performed to establish a regulated source of pulmonary blood flow. Most neonates undergoing a Starnes RV exclusion procedure are then channeled down a single ventricle pathway with a bidirectional Glenn and Fontan procedures.

The two competing strategies for surgical treatment for neonatal EA are whether to perform a biventricular repair or a single ventricle palliative procedure (Starnes Procedure).

The decision for which type of repair is best for neonatal EA is controversial. Mizuno et al. described their center experience with neonatal EA repair. Their results demonstrated greater survival for the biventricular repair group compared to single ventricle palliation, 60 vs. 25% respectively [30]. A recent follow up study by Kumar et al. evaluated the median 7 year follow up for the Starnes procedure in 27 neonatal repairs [29]. Their overall survival for 5 year follow up was 81%. Boston et al. described their outcomes for neonatal biventricular repair for EA. Early survival was 78.1% in their series, while 15 year survival for EA with and without anatomical pulmonary atresia was 40 and 79% respectively. As such caution should be advised for biventricular repairs in EA neonates with anatomical pulmonary atresia [25, 36].

In summary, neonatal EA continues to carry a high perinatal mortality upon fetal diagnosis. A multidisciplinary approach is required for improved outcomes. Fetal echocardiography predicts outcome and is necessary for monitoring progression of EA complications. Comprehensive care with a multi-disciplinary team including high risk obstetrician, pediatric cardiologist, pediatric cardiothoracic surgeon, neonatal intensivist should occur at a tertiary care center. Surgical management during the neonatal period remains high. If possible medical management through the neonatal period improves mortality.

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